QA 35 - Hydranencephaly QUESTION:

I have a client who is a 19 mos. old male with hydranencephaly. I have had difficulty researching this condition as I have found that there is not a lot of information regarding it. What is a normal developmental growth pattern for a child with this condition?

I am also currently working with the pediatrician to calculate this child's nutrition needs. Do you have any recommendations for calorie and protein needs for this child? He has been slipping on the NCHS growth chart and is less than the 5th percentile wt/age and at the 5th percentile wt/ht (a decline from his last visit). He has limited intake of baby foods secondary to developmental delays so nutrition supplements (PediaSure) are the base of his nutritional intake currently.

ANSWER:

Hydranencephaly is a major brain malformation where the cerebral hemispheres are absent and replaced by cerebrospinal fluid. It is very similar to anencephaly, except infants with hydranencephaly usually live longer. Sometimes hydrocephalus is also seen. Individuals usually have significant central nervous system impairment and developmental delays. Growth retardation is commonly seen.

Although the rate of growth may be slow and growth retardation is common, one would expect weight for length to be within the low normal to normal range. If motor development is very limited, muscle development is likely reduced. Therefore a lower weight/length (i.e. 5th-25th%) may be appropriate. Energy needs should be adjusted to improve wt/lgth status, and intake and growth monitored regularly. Protein intake would be typical based on size. Since many children with this diagnosis are severely delayed, use of a gastrostomy tube is common.

Reference:

1) Pipes, PL and Glass RP: Nutrition and Special Health Care Needs. In: Trahms, CM and Pipes, PL: Nutrition in Infancy and Childhood, 6th ed, 1997, WCB/McGraw-Hill.